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ORIGINAL ARTICLE

Uncommon congenital extracardiac vascular anomalies detected on MSCT (Multi-Slice Computed Tomography) aortic angiography with 64-multislice technology

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KEYWORDS

CTA;
Vascular;
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Abstract *Objective:* The purpose of this study was to evaluate the role of CT angiography in different encountered congenital vascular anomalies of pediatric age groups using 64 multislice CT scanner.

Subjective and methods: Through one year, a total of 15 patients having congenital extracardiac anomalies were encountered from those attending big trauma and emergency center and were studied in this work. Each patient's parent was thoroughly asked about the detailed clinical history after reviewing the referring imaging request and laboratory findings. Exclusion criteria for CT were as follows: previous allergic reactions to iodine contrast media and severe renal insufficiency. CT examination was performed using 64 multislice CT machine.

Results: CTA examination was performed in a total of 15 patients. The age of the patients was ranging from 2 days to 14 yrs and 5 cases were females. Four cases came with cyanosis; one case came with follow up after operation for transposition of great arteries, while the remaining cases were suffering from medical problems such as respiratory distress or hypertension. Aortic arch hypoplasia, right sided aortic arch with mirror image or with aberrant left subclavian artery, hypoplastic left pulmonary artery with left sided aortic arch and aberrant right subclavian artery, hypoplastic left heart chambers, retrotracheal left pulmonary artery with patent ductus arteriosus (PDA), right superior partial anomalous pulmonary venous drainage (PAPVD) with ASD sinus

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venosum, postoperative assessment of transposition of great arteries (TGA), fibromuscular dysplasia involving the renal and common iliac arteries, and bilateral double renal arteries with ectopic right kidney were the different extracardiac vascular anomalies encountered in this work.

Conclusion: CTA provides an excellent means to detect a number of extracardiac vascular anomalies and allows accurate and fast noninvasive characterization of extracardiac vascular anatomy. It is a helpful tool in establishing the primary diagnosis, defining anatomic landmarks and relationships, identifying vascular anomalies and helping in postoperative follow-up.

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1. Introduction

CT, including CT angiography (CTA), is important in the evaluation of pediatric congenital heart disease (CHD). It can be used for accurate depiction of complex cardiovascular anatomic features both before and after surgery and of a variety of post treatment complications. CT facilitates the assessment of extracardiac systemic and pulmonary arterial and venous structures. It is important for radiologists to have extensive knowledge of cardiovascular anatomy, physiology, and surgical techniques (1).

Pediatric thoracic CTA doses quoted in the literature vary greatly from 1–2 mSv (2,3) to more than 50 mSv (4). A variety of techniques have proved effective in lowering radiation exposure, including decreasing tube current (2,5,6) and potential (5,7,8), increasing pitch (6) and table speed (5,8), avoiding multiphasic imaging (5), and minimizing scan coverage. Rotational tube current modulation, a form of automatic exposure control, can be used to further lower radiation exposure (8–10).

CTA depicts the thoracic aorta noninvasively and in exquisite detail. Numerous congenital anomalies of the aorta can be diagnosed with this imaging technique, including anomalous arch branching patterns and configurations. CT also can be used to establish situs and L- versus D-looping of the great vessels (11). A combination of axial, 2D reformatted, and 3D reconstructed images can be used to accurately depict these anatomic relations (12).

CT depicts a number of congenital pulmonary arterial anomalies, including agenesis (13), hypoplasia (14), and proximal interruption (15,16). In pulmonary agenesis, CT reveals absence of the unilateral pulmonary artery and lung and ipsilateral mediastinal shift. A small unilateral pulmonary artery and lung and ipsilateral mediastinal shift are found in pulmonary hypoplasia. These anomalies may be isolated or associated with other cardiovascular anomalies (12).

2. Subjects and methods

2.1. Patient population

Through one year, a total of 15 patients having congenital extracardiac anomalies were encountered from those attending big trauma and emergency center and were retrospectively studied in this work. Each patient's parent was thoroughly asked about the detailed clinical history after reviewing the referring imaging request and laboratory findings. Exclusion criteria for CT were as follows: previous allergic reactions to iodine contrast media and severe renal insufficiency. CT examination was performed using 64 multislice CT machine. (As per

hospital policy, informed consent was taken and signed by the patient's parents and referring doctors.)

2.2. Patient preparation

- The patient was fast for about 2 to 3 hours before the appointment due to the introduction of contrast media in this examination.
- Hydration was recommended four hours prior to the exam.

2.3. Technique

2.3.1. Patient positioning and preparation for scanning

Patients are positioned on the CT examination table in the supine position. Intravenous access via a small intravenous line (e.g., 23 to 24 gauges) was necessary in neonates and infants and 20 gauge in older children to ensure easy injection of the viscous contrast agent. Manual injection was done in all cases.

A right arm injection was preferable to avoid artifacts from undiluted contrast media in the left brachiocephalic vein. A bolus of saline following the iodinated contrast media injection was done in some cases to reduce the volume of contrast media required to achieve adequate vascular opacification and reduce artifacts from high concentration of contrast media in the superior vena cava and right atrium.

2.4. Scan parameters

CT angiography protocol was created (Table 1). The protocol was dedicated to use the minimum parameters required to get acceptable diagnostic images with low radiation dose. Once patient preparation has completed, there are three steps to acquire data for the CT.

1. Scout scan
2. Contrast injection protocol
3. CT helical scanning

2.4.1. Scout scan

First two scouts with the patient in the head-first supine position were taken.

2.4.2. Contrast injection protocol

The administration of iodinated contrast media for the CT angiogram was done using non-ionic contrast media (omnipaque 300). The dose was 2 to 2.5 ml per kg. The contrast injection was done manually. After calculating the delay time

Table 1 Scan protocol.

Diagnostic protocol	
Application (scan type)	Helical
Region	Chest and abdomen
Kv	100–120
Effective ma	140–599
Scan coverage	Root of the neck to mid-abdomen /syphysis pubis
Rotation time	0.35 s
Collimation	64 × 0.625 mm
Slice thickness	0.625 mm
Slice interval	0.625 mm
Pitch	Set automatically
Recon interval	0.625 mm

to initiate CT scanning (It was between 1 to 3 seconds), I started contrast injection. After I have injected 60% to 70% of the total contrast dose, the radiographer started scanning while I was completing the remaining amount.

2.4.3. CTA scanning

This is the final scan before transferring the images to the workstation. When the scan was completed, I quickly checked the images. It should cover the complete chest and upper abdomen. The first (superior) image location should be at root of the neck. The last (inferior) image location should be below the kidneys. The whole chest and abdomen down to symphysis pubis was done in some cases, if the aorta was the primary site of interest.

2.5. Image evaluation

Axial images were transferred to the diagnostic workstation and were reconstructed for each patient and analyzed for motion artifacts. Both axial images and multiplanar reformatted (MPR) images were instrumental in detecting aortic abnormality. Although two-dimensional and three-dimensional reformatting techniques such as maximum intensity projection, curved planar reformation, multiplanar reformation, and volume rendering (VR) may facilitate interpretation and improve communication with referring physicians, axial images still remain the cornerstone of the evaluation, as virtually all pathologies can be recognized. Once a suggestive location with vascular abnormality has been identified on axial images, thin to thick-slab maximum-intensity-projection (MIP) images were created.

Double oblique reformatted images obtained perpendicular to the aortic lumen (i.e., true short-axis images of the aorta) allow more accurate measurement of aortic diameter than does relying on axial CT scans, on which the aorta has an oblique course (17). The direct measurement of lumen diameter was used at specific locations according to Agarwal et al. (18) (Fig 1).

The aortic and pulmonary diameter; the normal anatomy of the aorta and its great branches, pulmonary arteries and pulmonary veins; the origin of coronary arteries; assessment of ductus arteriosus, if present; and gross morphological anatomy were the target findings to be determined and emphasized on interpretation of CTA imaging.

Sinus
Sinoaortic junction
Midascending aorta (midpoint between sinoaortic junction and proximal aortic arch)
Proximal aortic arch (aorta at origin of brachiocephalic trunk)
Mid-aortic arch (just distal to left common carotid artery)
Proximal descending aorta (2 cm distal to left subclavian artery)
Mid-descending aorta
Aorta at diaphragm (2 cm above celiac axis origin)
Abdominal aorta at celiac axis origin
Abdominal aorta at most cephalic renal artery
Abdominal aorta at most caudal renal artery
Infrarenal abdominal aorta (15 mm below most caudal renal artery)
Aorta just above bifurcation
Aneurysm (maximum diameter [location specified])

Fig. 1 Specific location for aortic diameter measurements (18).

3. Results

3.1. Baseline clinical details (Table 2)

CTA examination was performed in a total of 15 patients. The age of the patients was ranging from 2 days to 14 yrs and 5 cases were females. Four cases had cyanosis; one case came with follow up after operation for transposition of great arteries, while the remaining cases were suffering from medical problems such as respiratory distress or hypertension.

3.2. CTA findings of different encountered extracardiac vascular anomalies

Aortic arch hypoplasia, right sided aortic arch with mirror image or with aberrant left subclavian artery, hypoplastic left pulmonary artery with left sided aortic arch and aberrant right subclavian artery, hypoplastic left heart chambers, retrotracheal left pulmonary artery with patent ductus arteriosus (PDA), right superior partial anomalous pulmonary venous drainage (PAPVD) with ASD sinus venosum, postoperative assessment of transposition of great arteries (TGA), fibromuscular dysplasia involving the renal and common iliac arteries, and double renal arteries bilateral with ectopic right kidney were the different extracardiac vascular anomalies encountered in this work (Table 3).

3.3. Aortic arch hypoplasia

One case with hypoplastic aortic arch was seen (Fig 2). He was 2 days old male. The whole arch was hypoplastic and the great arterial branches were coming from the hypoplastic segment with normal origins. The ascending and descending aorta appears of normal size and location. Patent ductus arteriosus was seen between the main pulmonary artery and aorta just distal to the left subclavian artery. It was about 8 mm length and 8 mm width. VSD was another associated anomaly.

3.4. Hypoplastic left pulmonary artery with aberrant Rt. subclavian

One case with hypoplastic left pulmonary artery was seen associated with normal morphological aortic anatomy and aberrant right subclavian artery (Fig 3). He was 9 weeks old. The

Table 2 Baseline clinical data.

Parameter	No. of patients(N:15)
Gender (M/F)	5/10
Range age	2 days to 14 years
Respiratory distress	8
cyanosis	4
hypertension	2
Postoperative follow up	1

Table 3 Different encountered vascular anomalies.

Vascular anomalies	Number
1. Aortic arch hypoplasia	1
2. Hypoplastic left pulmonary artery with aberrant Rt. subclavian	1
3. Right sided aortic arch	5
With mirror image type	4
With aberrant left subclavian artery	1
4. hypoplastic left heart chambers	1
5. Retrotracheal left pulmonary artery with PDA	2
6. PAPVD with ASD sinus venosum	2
7. Post-op for TGA	1
8. Fibromuscular dysplasia	1
9. Ectopic right kidney with bilateral double renal arteries	1

aberrant right subclavian artery was seen coming from the aorta distal to the left subclavian artery and passing through the retroesophageal space to the right side. Patent ductus

arteriosus (with 7 mm length and 5 mm width) was seen between the aorta distal to the origin of aberrant right subclavian artery and the hypoplastic left pulmonary artery with narrowed its pulmonary end.

3.5. Right sided aortic arch

Five cases with right sided aortic arch were encountered. Four cases showed mirror image type and aberrant left subclavian artery seen in the fifth one (vascular ring). Membranous ventricular septal defect (VSD) was seen in one case with mirror image type (Fig. 4). The remaining extracardiac vascular anatomy was normal.

3.6. Hypoplastic left heart chambers

One case with hypoplastic left atrial and left ventricular chambers was encountered (Fig 5). The ascending aorta was severely hypoplastic and seen coming from the hypoplastic left ventricle with hypoplastic aortic arch. The pulmonary artery was seen coming from the dilated right ventricle with muscular defect VSD seen between the two ventricles. Patent ductus arteriosus (PDA) was seen between the aorta distal to the hypoplastic arch and main pulmonary artery with likely retrograde filling of the hypoplastic arch and the normal size and origins of its great branches. The pulmonary veins were seen going to the small sized left atrium. ASD was also present. The coronary arteries were seen coming from the hypoplastic aortic sinus. The atrioventricular and aortic valves were not accurately assessed and believed to be atretic or hypoplastic. The provisional diagnosis was hypoplastic left heart syndrome. The

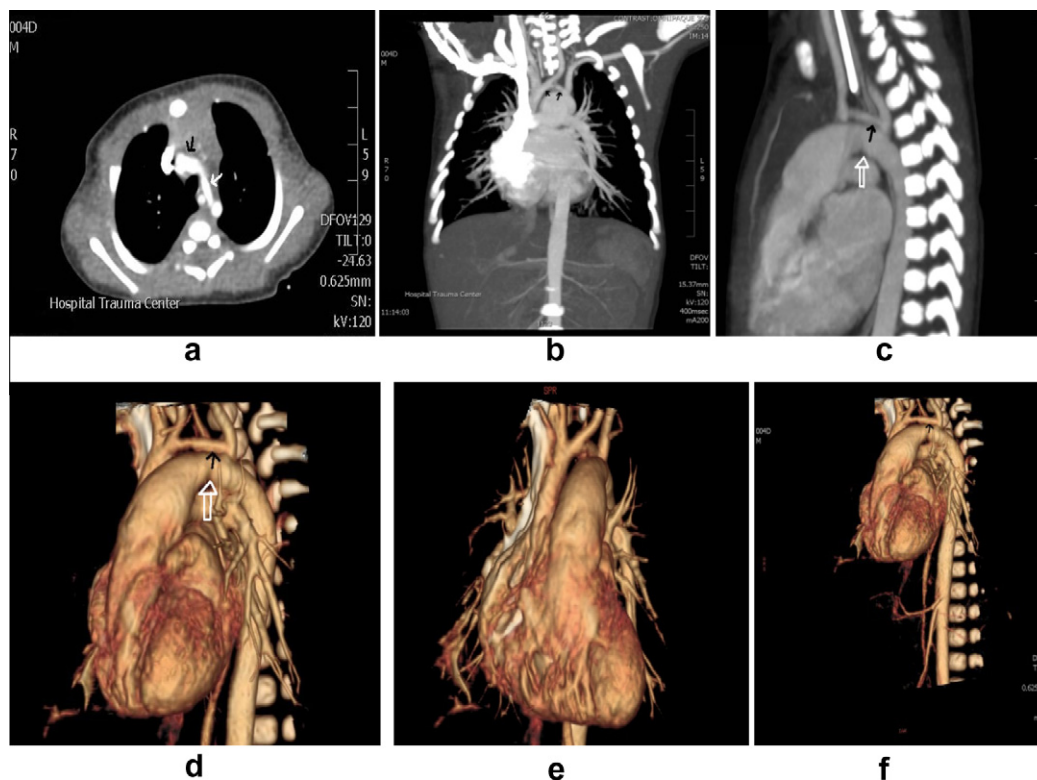


Fig. 2 Hypoplastic aortic arch. (a) Axial image, (b and c) coronal and sagittal MIP, and VR (d and e) shows: The whole arch appears hypoplastic (arrows) with patent ductus arteriosus between the pulmonary artery and aortic isthmus (open arrows in c and d).

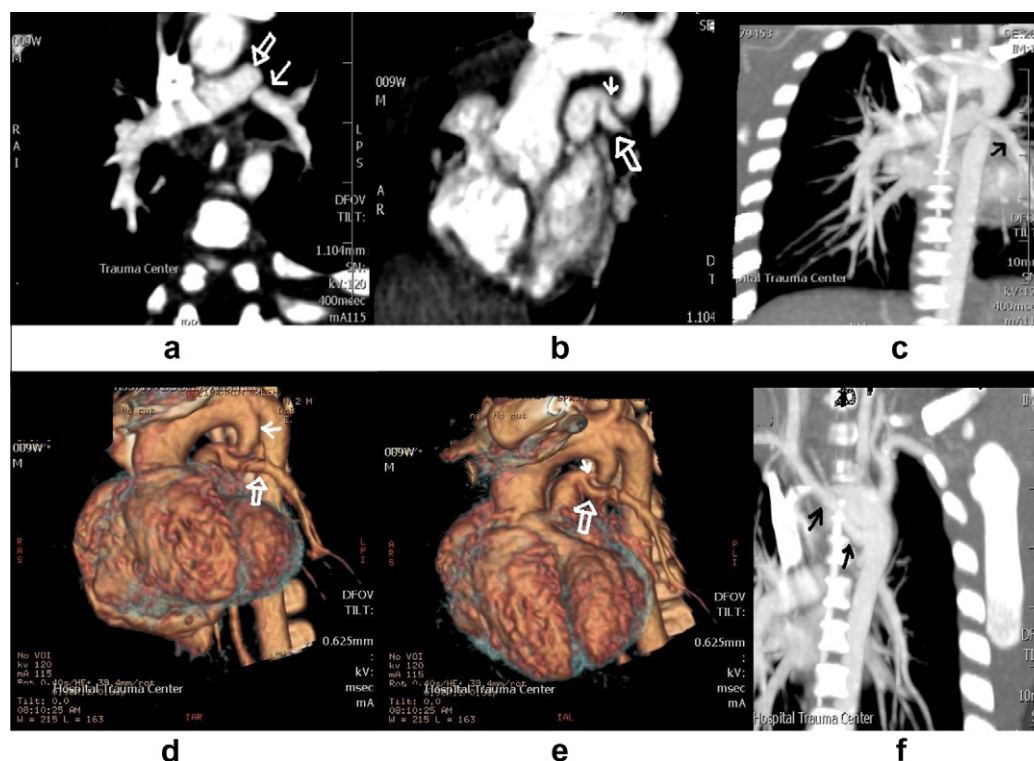


Fig. 3 Left pulmonary artery hypoplasia. Axial, sagittal, and coronal oblique MIP (a–c), VR (d and e) and coronal oblique MIP (f) images show hypoplastic, but normal course and origin, left pulmonary artery (arrows in a, c and open arrows in d and e). Patent ductus arteriosus is seen with narrowed pulmonary end (arrows in b and e). Aberrant right subclavian artery is incidentally seen (arrows in f).

final diagnosis has not been reached because I have lost the communication. (The patient was travelled abroad)

3.7. Retrotracheal left pulmonary artery with PDA

Two cases were encountered with anomalous course (retrotracheal) of left pulmonary artery but of normal size and shape (Fig 6). PDA was also seen in both cases. The first case was 4 months old boy while the second case was 2 months old female.

3.8. PAPVD with ASD sinus venosum

Two cases, already diagnosed as ASD, sinus venosus type, were transferred to the radiology department to assess the pulmonary veins drainage. Both cases showed partially anomalous pulmonary venous drainage. The right superior pulmonary vein was seen draining into the superior vena cava in both cases (Fig. 7).

3.9. Post-op for TGA

One case came to the radiology department for follow up for corrected transposition of great arteries. He was 14 years old. Successful correction was seen. The conduits between the pulmonary artery and right ventricles appear patent. Single coronary artery was seen coming from the aortic sinus (Fig. 8).

3.10. Fibromuscular dysplasia

One case was encountered with multiple segmental narrowing involving the middle and distal segments of right renal artery,

distal segment of left renal artery and both common iliac arteries (Fig. 9). She was 12 days old.

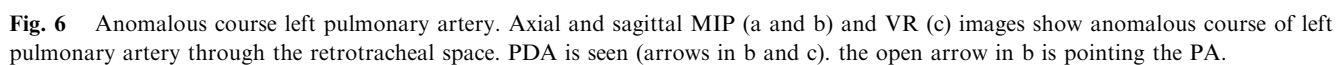
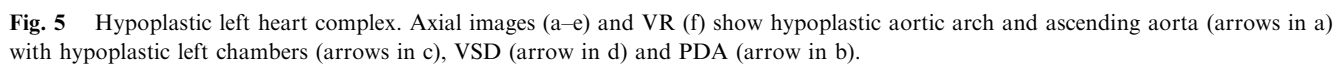
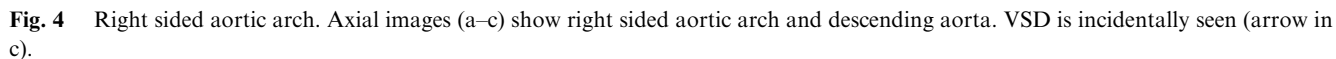
3.11. Ectopic right kidney with bilateral double renal arteries

One case came to the radiology department to rule out renal artery stenosis. The CT aortic angiography showed normal size and site of left kidney and left renal artery with accessory left renal artery coming from left common iliac artery and supplying the lower pole. The right kidney was ectopic seen in the right side of the pelvis with two renal arterial supplies. The first one was seen coming from the lower posterior aorta just above the bifurcation while the second one coming from the right common iliac artery (Fig 10).

4. Discussion

CT, including CT angiography (CTA), is important in the evaluation of pediatric congenital heart disease (CHD). It can be used for accurate depiction of complex cardiovascular anatomic features both before and after surgery and of a variety of post treatment complications. CT facilitates the assessment of extracardiac systemic and pulmonary arterial and venous structures. It is important for radiologists to have extensive knowledge of cardiovascular anatomy, physiology, and surgical techniques (1).

CT can be used to evaluate patients with congenital heart disease (CHD) known or suspected on the basis of echocardiographic findings for which further imaging is needed to characterize extracardiac anomalies before intervention. The pulmonary arteries, pulmonary veins, and aortic arch and



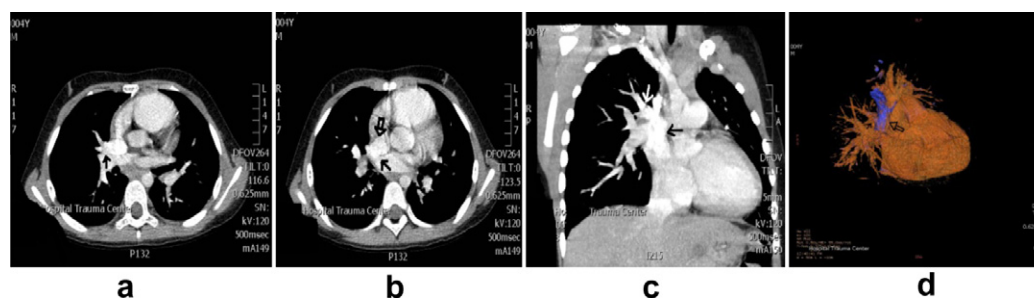


Fig. 7 Partially anomalous pulmonary venous drainage. Axial images (a and b), coronal MIP (c) and VR (d) show anomalous drainage of right superior pulmonary vein into superior vena cava (arrows in a and c, and open arrow in d). Sinus venosus defect is seen (arrow in b). The open arrow in b is pointing to part of right atrium.

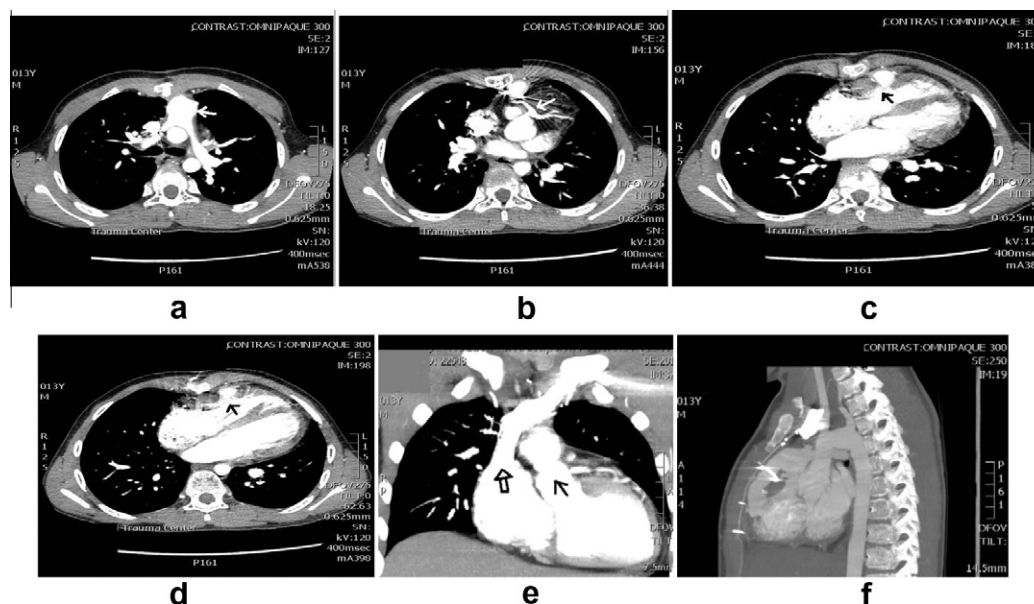


Fig. 8 Post operative follow up after correction of TGA. Axial (a–d) images and coronal e and sagittal f MIP show good arterial shift with anteriorly placed pulmonary artery (arrow in a). The conduit between the pulmonary artery and right ventricle appears patent (arrow in c and d). Single coronary artery is noted (arrow in b). The arrow in e is pointing to the aortic root while the open arrow is pointing to the IVC.

great vessels may be inadequately characterized at echocardiography, necessitating further assessment with CT (7).

In this work, a 64-multislice CT scanner was used to evaluate different encountered congenital extracardiac vascular anomalies in pediatric age groups. One case with postoperative evaluation was encountered. It was postoperative assessment after correction of TGA.

Pediatric thoracic CTA doses quoted in the literature vary greatly from 1–2 mSv (2,3) to more than 50 mSv (4). A variety of techniques have proved effective in lowering radiation exposure, including decreasing tube current (2,5,6) and potential (5,7,8), increasing pitch (6) and table speed (5,8), avoiding multiphase imaging (5), and minimizing scan coverage. Rotational tube current modulation, a form of automatic exposure control, can be used to further lower radiation exposure (8–10). In this study, dedicated protocol has been created for CT angiography with modulation of the pitch, KV, MA to get the best result with the least radiation exposure. The radiation dose was equal to CT chest and/or abdomen. Non-ECG gated CTA was used. The multiphase imaging has been avoided.

Gradual tapering of the distal aortic arch and isthmus is a normal finding in the first 3 months of life. Persistence of this stricture later on is pathologic. The proximal arch segment is defined as hypoplastic when its external diameter is less than 60% of that of the ascending aorta. The corresponding limit for hypoplasia in the distal arch is 50%, and for the isthmus the limit is 40%. The term tubular hypoplasia refers to a combination of abnormal small diameter and increased length (> 5 mm in infants) between the segments of the aortic arch. Aortic arch hypoplasia is frequently associated with other forms of obstruction or restriction to aortic flow, most commonly aortic coarctation. Other associated congenital heart anomalies may also be noted, including ventricular septal defect (VSD), atrial septal defect, and PDA (19). In this work, one case with aortic arch hypoplasia was encountered and it was of tubular type and associated with VSD and PDA.

Right aortic arch with aberrant left subclavian artery and left ligamentum arteriosum is the second most common vascular ring, accounting for 12–25% of cases. In this anomaly, the left subclavian artery arises from the descending aorta (often

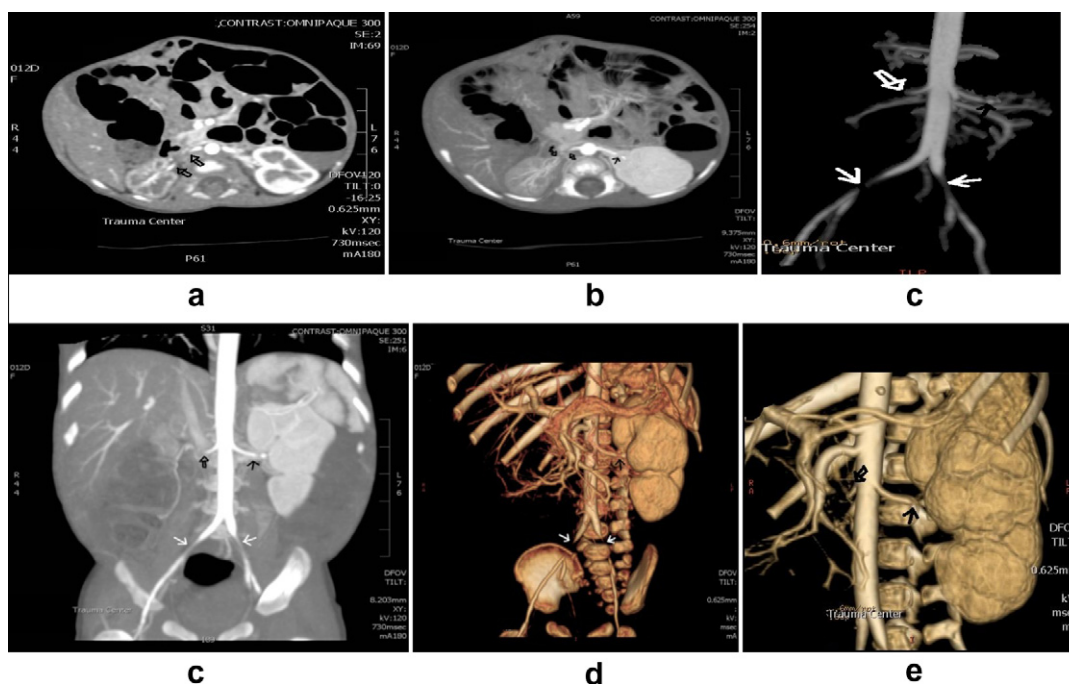


Fig. 9 Fibromuscular dysplasia. Axial cut and MIP (a and b), coronal MIP (c and d) and VR (d and e) show multiple segmental marked narrowing involving the middle segment right renal artery(open arrows), left renal distal segment (black arrows) and common iliac arteries (white arrows).

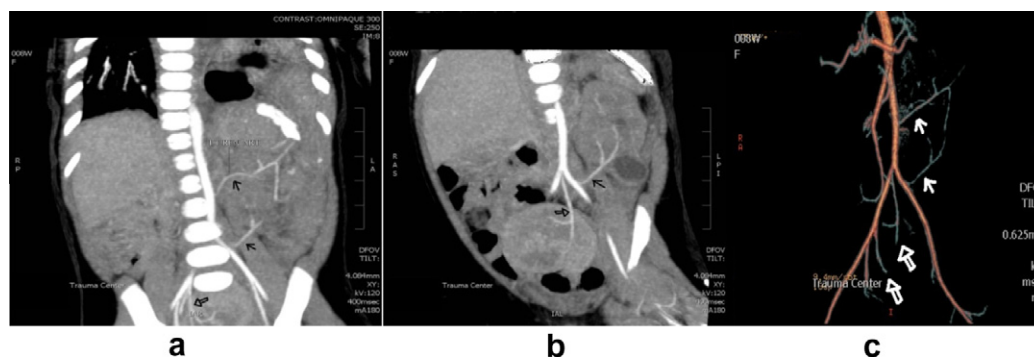


Fig. 10 Coronal MIP (a and b) and VR (c) show double bilateral renal arterial supply with ectopic pelvic right kidney.

from a diverticulum known as the Kommerell diverticulum) and runs posterior to the esophagus (20). These findings are seen in one case in this study.

CT can be used to characterize numerous pulmonary arterial congenital anomalies as well as to assess the postoperative appearance of the pulmonary arterial tree. CT facilitates accurate assessment of the central and peripheral pulmonary arteries (12). Anomalous origin of the left pulmonary artery from the proximal right pulmonary artery is referred to as a pulmonary sling and can be clearly visualized with CT (8,21,22). In this anomaly, the left pulmonary artery passes between the trachea and the esophagus on its way to the left pulmonary hilum (12). I have got three cases with pulmonary vascular anomalies, two cases with anomalous course left pulmonary artery, passing between the trachea and esophagus but of normal origin and size while the third one with hypoplastic left pulmonary artery with normal course and origin.

CT findings can be used to establish the diagnosis of partial and total anomalous pulmonary venous return in pediatric patients (2,7,8,23,24). In these conditions, the pulmonary veins drain to a location other than the left atrium. CT is a valuable noninvasive complement to echocardiography in the evaluation of pulmonary venous structures (25). In partial anomalous pulmonary venous return, at least one pulmonary vein drains normally to the left atrium. Partial anomalous pulmonary venous return most commonly involves drainage of right pulmonary veins to either a right SVC or the right atrium (8). Abnormal connection of the right upper pulmonary vein to the superior vena cava occurs frequently in the presence of a sinus venosus defect (26). In this work, two cases with partially anomalous pulmonary venous drainage were seen; both of them showed anomalous draining of right superior pulmonary vein into superior vena cava and associated with ASD, sinus venosus type.

In patients with a borderline small ventricle (as in hypoplastic left heart complex or an unbalanced AVSD, the size of the small ventricle determines prognosis and is an important parameter for deciding between biventricular repair and univentricular palliation (27). One case with hypoplastic left atrium and ventricle was encountered with VSD and probably ASD with hypoplastic ascending aorta and aortic arch with likely retrograde contrast opacification from PDA.

Today, surgical repair is usually performed in transposition of great arteries by using the arterial switch procedure, which has replaced both the Senning and the Mustard atrial switch operations. In the arterial switch operation, the aorta and the pulmonary artery are transected above the valves and moved to the correct circulatory position. The coronary arteries are excised from the right side with a button like margin of tissue around each artery and are implanted just above the valve on the left side of the heart; the areas from which the coronary arteries were excised then are patched with pericardium (7). I have got one case with postoperative follow up for correction of transposition of great arteries. It was arterial shift operation with successful results. Single coronary artery was seen coming from the aorta.

Fibromuscular dysplasia is a less frequent cause of renovascular hypertension and is more frequently seen in women at a younger age; it manifests as a string-of-beads appearance (28). I have got one case with fibromuscular dysplasia with right middle and left renal stenosis associated with high grade bilateral common iliac stenosis. She was 12 days old and came with renovascular hypertension.

5. Conclusion

CTA provides an excellent means to detect a number of extracardiac vascular anomalies and allows accurate and fast non-invasive characterization of extracardiac vascular anatomy. It is a helpful tool in establishing the primary diagnosis, defining anatomic landmarks and relationships, identifying vascular anomalies and helping in postoperative follow-up.

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